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CLINICAL CASE.

HEREDITARY CASES OF PROGRESSIVE MUSCULAR ATROPHY.

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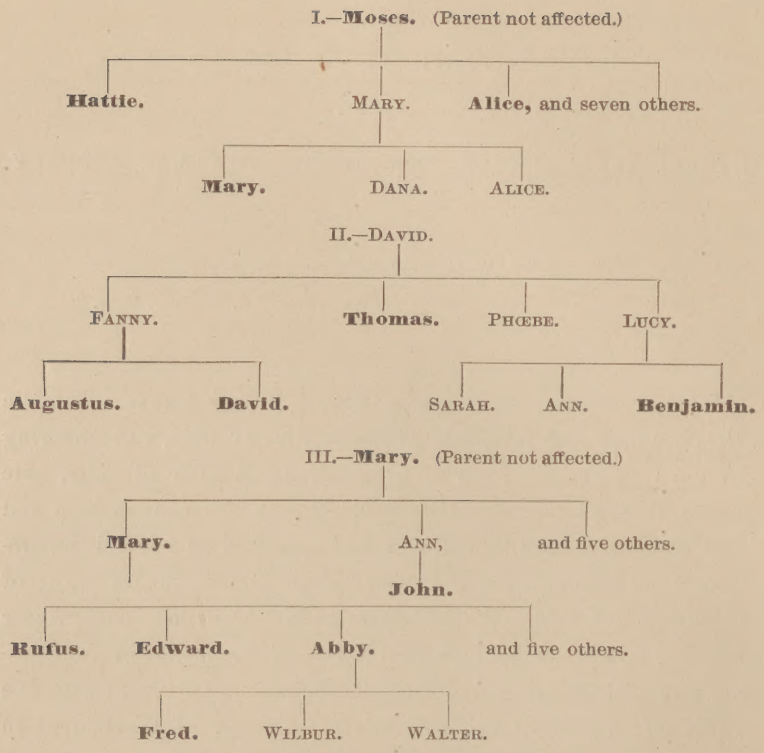
In February last there was admitted to the Danvers Lunatic Hospital a patient who, aside from mental trouble, was suffering from a very chronic form of progressive muscular atrophy. He stated that he had relatives afflicted with the same malady and that there were traditions that it had appeared in several generations upon his mother's side. Surmising that this might be one of those families, a few of which have been placed on record, where heredity appears to have exerted a powerful influence in propagating the disease for generations, an inquiry was begun with the purpose of verifying the patient's statements. This resulted in bringing to light fifteen well-authenticated cases, all springing from a common ancestry and extending over a period of about one hundred and fifty years. How many other cases there have been it is impossible to ascertain. Of some, traces have been obtained here and there, but of so indefinite a character that they have not been included in this account.

The following *schema* will readily show the manner in which transmission has occurred. It has been found convenient to arrange them in three groups. Moses, the head of group I, is a direct descendant of one of two brothers; David and Mary, the respective heads of groups II and III, are direct descendants of the other brother, the said Mary being the paternal aunt of the said David. The names in heavy type represent the individuals affected:



From the above we are able to deduct the following facts. Of these fifteen cases nine are males and six females. They are the offspring of nine marriages. Transmission through the females to children has occurred in eleven cases, eight being males, through the male to children in four cases, three being females. Where the mother herself has been affected, the disease has been transmitted to children five times, where the father has been affected, twice. There are six cases whose parents were not affected.

It is said that the male sex shows a much greater tendency to the disease than the female. Out of one hundred and seventy-six cases collected by Friedreich only thirty-three were females. Another peculiarity is for the disease to be transmitted through the female rather than through the male as shown in the cases described by Hemptenmacher. While the cases being cited present exceptions, yet it may be seen that in the majority of instances they conform to these two principles.

Trousseau mentions a family in which the great-grandfather, grandfather, father and son suffered from the disease. A still more striking example of atavism is presented in group III where it will be observed that a mother, one child, four grandchildren and one great-grandchild are all affected.

Eight of these fifteen cases are living. I have visited those within my reach, inquired into their history and made a personal examination when permitted. The following is a brief account of a few of these cases:

Group I—Lydia, æt. forty-nine, dressmaker, when thirty-five years of age, first began to notice the affection in her feet. At present there is marked atrophy of all the muscles of the feet with flexion of the toes. The muscles of the lower legs are atrophied, but in a less degree. As yet there have been no symptoms connected with the upper extremities.

Alice, sister of the preceding, died, æt. fifty-three, of pulmonary disease. She began to be affected in the lower extremities at about the age of thirty-five. During the last five years of her life, her upper extremities began to show the disease.

Group II—David, æt. sixty-five, muscles of the hands and feet much atrophied, fingers and toes contracted in flexion. Began to be troubled before he was twenty. Has followed the occupation of teamster nearly all his life.

Group III—Edward, æt. fifty-two, referred to as the patient now in the hospital. States that his trouble began at about fifteen years of age, and nearly simultaneously in the upper and lower extremities. For a few years the disease advanced pretty rapidly, but since the age of thirty-five, there has been no marked progress unless it is increasing contractions. At present there is atrophy of all the muscles of the hands and fingers, with a lower degree of wasting of the fore-arm. There is a similar condition of the feet and legs. For the greater part of his life he has been a stitcher on shoes; his last occupation was that of a peddler of small wares.

The history and present condition of his brother Rufus and sister Abby correspond so closely to his own that they would be essentially repetitions. They both began to be affected at about the age of eighteen. In Abby's case the affection appeared in her feet nearly two years before it showed itself in the hands. Rufus is a house-painter, and is still carrying on his occupation. Abby is able to do her housework.

Fred, æt. twenty-nine, sandpaperer, began to be affected in the

feet when eighteen years old, and a year later symptoms appeared in his hands. To-day his upper and lower extremities are affected nearly as much as in the three preceding cases.

In this collection of cases, so far as I can learn, the disease has almost always appeared before the age of twenty, has advanced up to a certain point, after which its progress has been extremely slow. In no instance can I find that parts above the elbow or knee have been invaded to any extent, and no death has been attributed to this affection. The general health does not appear to suffer in any way, neither does longevity seem to be affected. One of these cases, it is said, lived to be a centenarian. Some of them have reared large families of children; they have all been able to indulge in some form of manual labor, by which a livelihood has been obtained, and I have yet to learn that one of them became a pensioner upon his friends until late in life, when causes connected with senility were more accountable than the life-long disease.

Edward is the only instance of insanity in the family within the recollection of his friends. The healthy members are robust-looking individuals, and the family history does not reveal any other neurosis. A marked contrast is presented between the course of the disease in this family and that in the Wetherbee family of Western Massachusetts, reported by Dr. W. A. Hammond. In the latter the disease did not appear until the subjects were well advanced in adult life, when its progress was comparatively rapid, ending fatally in a few years.

These cases are very useful as showing the fact that progressive muscular atrophy of *hereditary* origin is not necessarily a fatal malady; that it may pursue an extremely chronic course; that, so far as general health and longevity are concerned, it may be innocuous, and finally, that, after advancing to a certain point, it may lose essentially its *progressive* character.

I feel that I should be betraying a confidence were I to publish the names of these people in full. I therefore represent the family name of their common ancestry as follows: P******. Should cases springing from this same source be observed by others, I think that the name would be readily recognized. The people who have formed the subject of this paper are inhabitants of Eastern Massachusetts. The collateral branches are numerous and are found in every part of the country.

I am indebted for family data in this paper to a carefully prepared genealogy, published a few years ago, in the absence of which I could not have traced these cases to a common parentage.